SYDENHAMS CHOREA AND INSIDIOUS CARDITIS IN A - 9 YEAR OLD GIRL

Putu Ayu Widyanti, Eka Gunawijaya, Dewi Sutriani
Department of Child Health, Medical School, Udayana University/ Sanglah Hospital, Denpasar

ABSTRACT

Sydenham's chorea is the most common form of acquired childhood chorea, and represent one of the major diagnostic criteria of rheumatic fever. It characterized by involuntary movements which disappear during sleep, emotional instability, and hypotonia. We presented a - 9 year old girl with choreatic movement (involuntary movement of arm and feet). The unwanted movement also found on tounge made difficult to speak. She also had emotional lability, and muscle weakness. The history of trauma was denial. On physical examination found murmur at apex area, holosistolic, grade 2/6, blowing, and spreading along axilla. On echocardiography found moderate mitral regurgitation (MR) and trivial aortic regurgitation (AR) due to carditis. The diagnosis was sydenham's chorea and carditis. She was treated with erythomicin 250 mg four times a day for 10 full days, and followed by erythromycin 250 mg orally twice a day for prophylaxis. For symptomatic treatment, haloperidol 2 mg twice a day and trihexyphenidil 0.5 mg three times a day was given. The therapy respon and prognosis was good. **(MEDICINA 2012;43:54-59).**

Keywords: *Sydenham's chorea, rheumatic fever, carditis*

KOREA SYDENHAM DAN KARDITIS TERSEMBUNYI PADA SEORANG ANAK PEREMPUAN USIA 9 TAHUN

Putu Ayu Widyanti, Eka Gunawijaya, Dewi Sutriani
Bagian/ SMF Ilmu Kesehatan Anak Fakultas Kedoteran Universitas Udayana/ Rumah Sakit Umum Pusat Sanglah, Denpasar

ABSTRAK

Korea *Sydenham* adalah bentuk paling umum dari gerakan korea yang diperoleh pada masa kanak-kanak, dan merupakan salah satu kriteria diagnostik utama demam rematik. Korea *Sydenham* ditandai dengan gerakan involunter yang menghilang saat tidur, ketidakstabilan emosional, dan hipotonia. Kasus adalah anak perempuan berumur 9 tahun dengan gerakan *coreatic* (gerakan involunter pada lengan dan kaki). Gerakan tersebut juga ditemukan pada lidah sehingga pasien sulit untuk berbicara. Pasien juga memiliki ketidakstabilan emosi, dan kelemahan otot. Riwayat trauma disangkal. Pada pemeriksaan fisik ditemukan murmur di daerah apex jantung, holosistolik, derajat 2/6, meniup, dan menyebar sepanjang aksila. Pada *echocardiography* didapatkan regurgitasi mitral moderat (MR) dan regurgitasi aorta (AR) karena karditis. Diagnosisnya adalah Korea *Sydenham* dan karditis. Pasien diterapi dengan erythomicin 250 mg empat kali sehari selama 10 hari, dan eritromisin 250 mg oral dua kali sehari untuk profilaksis. Untuk terapi simtomatik diberikan haloperidol 2 mg dua kali sehari dan trihexyphenidil 0.5 mg tiga kali sehari. Respon terapi dan prognosis baik. **(MEDICINA 2012;43:54-59).**

Kata kunci : *korea Sydenham, demam rematik, karditis*

INTRODUCTION

Sydenham's chorea is the most common form of acquired childhood chorea, and represents one of the major diagnostic criteria of rheumatic fever, aproximately in 10-30% of patients with rheumatic fever. Sydenham's chorea is caused by antibodies against group A β-hemolytic streptococcus bacteria, which cross react with the basal ganglia.[1-4] The majority of patients with Sydenham's chorea is characterized by unwanted nonstereotypedchoreatic movements, which disappear during sleep. One common sign is motor impersistence, which can be demonstrated by an inability to sustain eye closure or tounge protrusion. Other associated signs include grimacing, clumsiness, dysarthria, difficulty in dressing, writing and feeding, muscle weakness, and hypotonia.[1] The majority of patients with Sydenham's chorea present at age 5-15 years, with a female preponderance.[4]

In contrast to other manifestations of rheumatic fever, such as arthritis and carditis , which emerge 1-3 weeks after group A β – hemolytic streptococcus infection. Sydenham's chorea can present up to several months after infection. Sydenham's chorea is accompanied by carditis in 40-80% of patients. In 20-70% of patients, chorea is the only manifestation.[1]

Because of improved socioeconomic and sanitary conditions, along with the

widespread use of penicillin, the incidence of both Sydenham's chorea and rheumatic fever has declined in developed countries. But some cases still occur. Familiarity with Sydenham's chorea is therefore required for physicians, especially because Sydenham's chorea can be a marker of life threatening carditis which led to write this case reporting.

THE CASE

KM, a 9-year-old-girl, has been refered to Sanglah Hospital by Bangli Hospital, with diagnosis of suspected space occupying lession. The major complain were uncontrol movement of arm and feet, just like people dance, since 1 week before admitted when she was at school. It occured suddenly and made she couldn't write and speak properly. There were no fever, no cough nor runny nose at that time. The uncontrolled movement disturbing her daily activity, and it stopped when she were sleeping.

The history of trauma was denial, history of palpitation also denial. There were no history of pain of the joint neither sore throat. Patient had normal passage of the stools and urine. There was no history of consuming medication such as antivomiting, antihistamine nor anti-epileptic drug.

She was born full term, spontaneously, and was assisted by her father. He didn't know about her birthweight and birthlength. No visible abnormalities were found. She did not get any prior medications for specific diseases. Her immunization status was complete according to the recommended immunization plan by government. She had normal milestones. Now she is at 4th grade elementary school. Daily activities prior to illness were normal. She participated well in school and in peer activities after school time. There was no history about family with the same complaint.

Physical examination revealed an alert girl. The blood pressure was 100/70 mmHg. The pulse rate was 98 x/min and regular, respiratory rate 28 x/min, axillary temperature 36.9°C. Her body weight was 26 kg, body height 129 cm. Her ideal bodyweight was 26 kg. Thus, according to the Waterlow criteria, her nutritional status was wellnourished (100% standard).

The hair was fine and black. The conjunctiva were not looked anemic, the sclera were not looked icteric, and the pupil reflexes were normal. The ear, nose, and throat as well as neck examination were within normal limit. There were no palpable lymph nodes or nuchal rigidity. The chest examination revealed in inspection we found no precordial bulging, ictus cordis was not appeared but palpable at intercostal space (ICS) IV left midclavicular line (MCL), right ventricle heave were not found, and there was no thrill. Heart beat sound was regular, with murmur at apex area (left ICS IV MCL), grade 2/6, blowing, holosistolic, and spreading along axilla. The movement of both sides of the chest was symmetrical. Vesicular respiratory sounds were heard, without wheezing or rales.

The examination of abdomen, bowel sound was normal, there were no hepatomegaly and splenomegaly. The examination of upper and lower extremities showed no deformities, no edema, and no cyanosis. The physiological reflexes of the patella and achilles tendon were normal, there were not found pathologic reflexes. Motor strength of the four extremities were normal.

Investigation of the chest X-ray revealed normal with cardio-thoracic ratio 49%, left aortic arch. The lung showed normal pulmonary vascular markings and there was no infiltrate (**Figure 1**). The electrocardiograph features revealed heart rate 83 times/minute with sinus rhythm, normal axis deviation and PR interval (**Figure 2**). Echocardiograph revealed atrial situs solitus, normal systemic and pulmonal veins drain, AV-VA concordant, balance 4 chambers, no atrial septal defect (ASD), ventricle septal defect (VSD), patent ductus arteriosus (PDA), nor coarctasio aorta. Moderate MR (ERO 0,1 cm2, R Vol 16 mL), trivial AR, normal pulmonal & tricuspid valves, left Ao arch, no pericardial effusion, normal LV systolic function (EF 80%). Echocardiograph conclusion : moderate MR & trivial AR due to carditis, which can seen in **Figure 3.** The head CT scan revealed normal **(Figure 4).** The CBC revealed WBC:17.2 k/μL (Ne 86.0%; Ly 12.7%), HGB 15.1 g/dL, HCT 43.5 %, MCV 79.9 fL, MCH 27.8 pg, PLT 248 k/μL. Serum natrium 136.20 mmol/L, serum potassium 4.172 mmol/L, chloride 98.73 mmol/L, calcium 9.697 mg/dL. Blood glucose 116 mg/dL. Anti-streptolisin titer O (ASTO) 400 IU/ml. C-reactive protein (CRP) 0.505, erythrocyte sedimentation rate (ESR) 10 mm. We also examined throat swab and the results were coccus gram positive and coccus gram negative, but the culture of the throat swab revealed Enterobacter sakazakii.

Based on the clinical manifestation, imaging finding, and laboratory results, we assessed the patient with rheumatic chorea (Sydenham's chorea) and insidious onset rheumatic carditis. The management to the patient were diminished of the activity and emotional stress, bed rest for 3-4 weeks, erythromycin 250 mg four times daily for 10 full days, haloperidol 2 mg twice daily, trihexylphenidil 0.5 mg three times daily, aspirin 650 mg, four times daily for 2 weeks, then we tapering

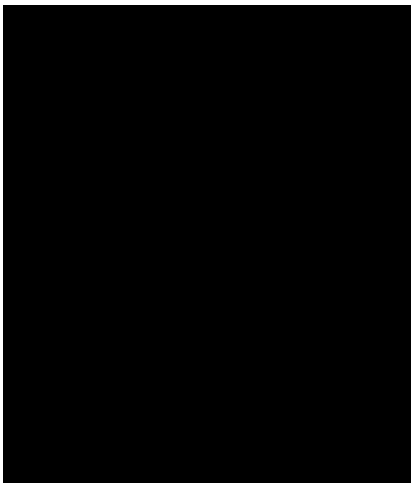

Figure 1. Normal plain chest X-ray was taken at the first admission.

Figure 2. Electrocardiogram, no prolongation of P-R interval.

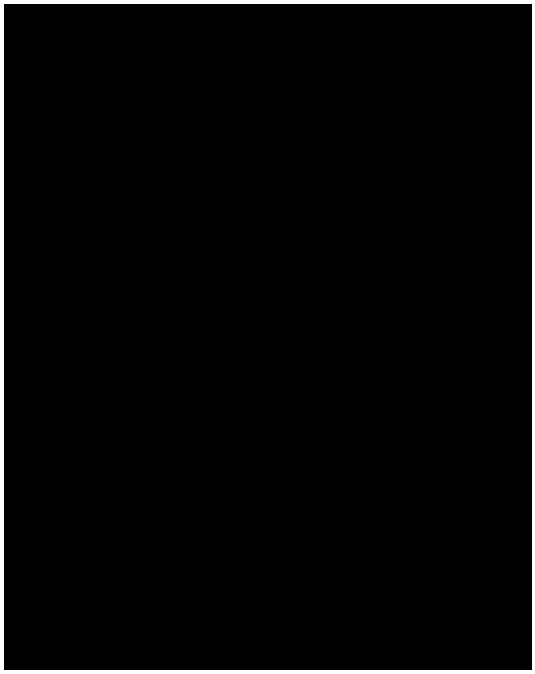

Figure 3. Echocardiography: moderate MR & trivial AR due to carditis.

Figure 4. The CT scan revealed normal.

off for 2-3 weeks. Long term management were secondary prophylaxis, we gave erythromycin 250 mg orally twice daily.

The patient's condition after treatment was improved. The involuntary movement decrease slowly and totally disappeared on 9th day after admitted, but patient still have to stay until 3 weeks more due to treatment of carditis. Patient was discharge at September 21st 2010 (11 days after admitted), without doctor recommendation, because of financial problem. We gave her erythromycin prophylaxis 250 mg orally twice daily, and suggestion to diminish her activity and get rest until next 3 weeks. We recommended to the patient to control to Sanglah hospital cardiology outpatient clinic and ASTO will retest again 2-3 months later.

DISCUSSION

Sydenham chorea, is the most common form of acquired childhood chorea, and represent one of the major diagnostic criteria of rheumatic fever, caused by anti-basal ganglia antibodies from a group A β hemolytic streptococcus infection. One common sign is motor impersistence, which can be demonstrated by an inability to sustain eye closure or tounge protrusion. Other associated sign include grimacing, clumsiness, dysarthria, difficulty in dressing, writing, and feeding, muscle weakness, and hypotonia.[1,4,5]

Because of the vulnerability of the basal ganglia and its connection to a wide variety of pathologies, the differential diagnosis of acute and chronic chorea is very large. Genetic and metabolic diseases, endocrine disturbances, autoimun disorders, infection, cerebrovasculer diseases, neoplasm, neurodegenerative diseases, toxins and trauma can all results in chorea.[4] The differential diagnosis of choreic syndromes relies not so much on differences in the phenomenology of the hyperkinesia but the presence of accompanying

findings.[4]

Most patient with sydenham's chorea have other symptoms of rheumatic fever. Sixty-80% of patient have cardiac involvement, particularly mitral valve dysfunction, whereas arthritis is present in 30 % patients; however, in about 20% of patients, chorea is the only finding.[4] In our case, chorea presented with carditis.

The majority of patients with sydenham's chorea present at the age 5-15 years, with a female preponderance.[1] In our case was a girl 9 years old, with unwanted nonstereotyped choreatic movements, which disappear during sleep. The unwanted movements like dancing, made an inability to dressing, writing, feeding, speaking, emotional lability, and made muscle weakness.

The perfectness of the skeleton muscle motoric function, needs a collaboration between pyramidal system and extrapyramidal system. Pyramidal system for the voluntary movement, and extrapyramidal system to control motoric impuls and consist of [1]. cerebri cortex nucleus area 4s, 6 and 8); [2]. nucleus of basal ganglia; [3]. Ruber nucleus and formation reticularis; [4].cerebelum. There are 3 circuit pathways for motoric impuls processing:[6] [1].the first circuit begin from motoric impuls which trigerred on area 4 and 6, delivered into basal pons nucleus, cerebellum cortex, dentatus nucleus, ruber and ventrolateral nucleus, and finally back to motoric cortex [2]. second circuit which passed by cerebri cortex area 4,4s and 6 going to substantia nigra, putamen, globus pallidus, ventrolateralis talami nucleus, and going back to motoric cortex [3]. third circuit motoric impuls from area 4s and 8 will pass trough this way, going to caudatus nucleus, globus pallidus, and ventrolateralistalami nucleus, and going back to cortex motoric area.

If there are a disturbance on one of the circuit or basalis ganglia nucleus or cerebellum, due to disturbance of feed back in to the motoric cortex area. Thus make an uncontrolled movement or we called involunter movement.

Chorea may develop following the other manifestation of acute rheumatic fever, or as an isolated neurologic disorder. As the latent period between the preceding infection and the development of chorea is prolonged (1-6 months), the usual supportive laboratory findings (elevation of acute phase reactans and streptococcal antibodies) may have returned to normal by that time.[1,2] A throat culture can confirm a preceeding group A βhemolytic streptococcus infection. However this culture is positive in only a minority of cases (< 15%). More often, serum analysis can demonstrated an elevation of erythrocyte sedimentation rate, of the C- reactive protein level. The most commonly performed and commercially available test are the the antistreptolysin – O and antideoxyribonuclease B test. The blood titres of antistreptolysin – O and antideoxyribonuclease B raised against extracellular antigens of streptococci reach a peak 3-4 weeks after the onset of group A β hemolytic streptococcus pharyngitis, and maintained for 2-3 months before declining. The presence of anti-basal ganglia antibodies in cerebrospinal fluid can be determined using Western immunobloting.[2] In our case, the history of infection at the throat was not clear, and chorea was accompanied with carditis without cardiomegaly, with normal acute phase reactans, and increase of ASTO titer. The anti basal ganglia antibodies can't be revealed.

Hospital admission may be helpful for confirming diagnosis of rheumatic fever, for instituting treatment and for educating the patient and family. All patient with acute rheumatic fever should be placed on bed-chair rest and monitored closely for the onset of carditis. In patient with carditis, a rest period of at least four weeks is recommended although physicians should make this decision on individual basis. Patient with chorea must be placed in a protective environment so they do not injure themselves. Majority studies described a treatment regime in three parts: (1) treatment of the underlying infection, (2) prophylaxis, and (3) symptomatic treatment.[2]

For the underlying infection, group *A β-hemolytic Streptococcus* infection, treatment of antibiotic is effective even most patient no longer harbor a symptomatic pharyngitis by the time the chorea becomes evident. A full 10-day course of oral penicillin V therapy or an injection of benzathine penicillin G provides effective treatment for patients with symptomatic or nonsymptomatic group *A β-hemolytic Streptococcus* infection (**Table 1**). For patients with allergy to penicillin, the macrolide erythromycin has been the recommended antibiotic of choice.[1,2] In our case the treatment of underlying infection is erythomicin 250 mg four times a day for 10 full days.

Antibiotic prophylaxis or secondary prevention of rheumatic fever (RF) is defined as the continuous administration of specific antibiotics to patient with a previous attack of RF, or well documented rheumatic heart diseases (RHD). The purpose is to prevent colonization or infection or upper respiratory tract (URT) with group A β-hemolytic Streptococcus infection and the development of recurrent attacks of RF four weeks, because of consistent findings.

The World Health Organization formulated indications for the duration of prophylaxis, depending on the patient's cardiac condition (**Table 2**).[2] In our case, the patient got erythromycin 250 mg orally twice a day.

For management of chorea often requires no specific treatment, because in most cases it is a benign, self limiting condition with a mean duration of 2-4 months. However, for patients in whom the chorea is debilitating and protracted, treatment is necessary. Neuroleptic, benzodiazepines and antiepileptics are indicated, in combination with supportive measures such as rest in a quiet room. Haloperidol, diazepam, carbamazepine have all been reported to be effective in treatment of chorea.[2] The dose of haloperidol is 0.01 mg/kg (max 0.5 mg) daily, increase up to 0.1 mg/kg 12 hour intravein or oral up to 2 mg/kg (max 100 mg) 12 hours used rarely. To prevent the side effect of haloperidol, add trihexyphenidil 0.02 mg/kg every 8 hours, increase to 0.1-0.3 mg/kg. In our case, for the management of chorea we give haloperidol 2 mg twice a day, and to minimizing the side effect of haloperidol, extra pyramidal effect, we gave trihexyphenidil 0.5 mg three times a day. She response to haloperidol quite quickly and theres were no observable side effect.

The antiinflamatory drugs that ussualy use are salicylates or corticosteroid. There is no convincing evidence in the literature that steroids are beneficial for the therapy of the chorea associated with rheumatic fever. We gave aspirin 100mg/kg-day divided into 4-5 doses. After achieving the desired initial state-steady concentration for two weeks, the dosage can be decreased to 60-70 mg/kg-day for an additional 3-6 weeks. We have to see the clinical response and normalization of acute phase reactans.[2] In our case for the antiinflamation we gave aspirin 650 mg, four times daily for 2 weeks, then we tapering off for 2-3 weeks. The prognosis of the patient was good, because the jerky movement disappeared after 9 days since hospiulaized.

Table 1. Therapeutic recommendations of the American Heart Association for the primary prevention of rheumatic fever in the treatment of GABHS tonsillopharyngitis

Agent	Dose	Mode	Duration
Benzathine penicillin G			
Patients <27 kg (60 lb)	600,000 units	IM	Once
Patients >27 kg (60 lb)	1,200,000 units	IM	Once
Penicillin V	children: 250 mg 2-3 times daily	PO	10 d
(phenoxymethyl penicillin)	adolescents and adult: 500 mg 2-3 times daily	PO	10 d
Erythromycin	20-40 mg/kg/d; divided 2-4 times daily (maximum 1 g daily)	PO	10 d
Estolate			
or		PO	
Ethylsuccinate	40 mg/kg/d; divided 2-4 times daily (maximum 1 g daily)		10 d

Table. 2 Antibiotics used in secondary prophylaxis of rheumatic fever

Antibiotic	Mode of administration	Dose
Benzathine benzylpenicillin	Single intramuscular injection every 3-4 weeks	For adults and children ≥30kg in weight: 1,200,000 units For children <30 kg in weight:600,000 units
Penicillin V	Oral	250 mg twice daily
Sulfonamide (e.g. sulfadiazine, sulfadoxine, sulfisoxazole).	Oral	For adults and children ≥30kg in weight: 1 gram daily For children <30 kg in weight: 500 mg daily
Erythromycin	Oral	250 mg twice daily

SUMMARY

A - 9 year old girl with choreatic movement (involuntary movement of arm and feet), difficult to speak, emotional lability, and muscle weakness was reported. Based on anamnesis, physical and laboratory examination, the patient was diagnosed with Sydenham's chorea. The treatment of underlying infection was erythomicin 250 mg four times a day for 10 full days, for prophylaxis was erythromycin 250 mg orally twice a day, and for symptomatic treatment was haloperidol 2 mg twice a day. We gave trihexyphenidil 0.5 mg three times a day to minimize the side effect of haloperidol, extra pyramidal effect. The therapy respon was good, involuntary movement decrease, and totally disappeared on 9th day after therapy. The prognosis also good.

REFERENCES

1. Oosterveer DM, Plandsoen WO, Roos RAC. Sydenham's chorea: A practical overview of the current literature. Pediatr Neurol. 2010;43:1-6.
2. Rheumatic fever and rheumatic heart diseases. Geneva: WHO; 2004.
3. Gilbert DL. Acute and chronic chorea in childhood. Seminar in pediatric neurology; 2009 [cited 2010 september 25th]. Available from:http:// www.elsevier .com
4. Cardoso F, Seppi K, Mair KJ, Wenning GK, Poewe W. Seminar on choreas. Lancetneurol. 2006;5:589-602.
5. Carepetis JR, McDonald M, Wilson NJ. Acute rheumatic fever. Lancetneurol. 2005; 366;155-68.
6. Rolak LA. Basal ganglia and movement disorder. In:Hanley, Belfus, editors. Neurology secrets. 3rd edition. Philadelpia: Hanley & Belfus Inc.,2001; p. 147-8.
7. Abdo WF. The clinical approach to movement disorders; 2010 [cited 2010 24th October]. Available from:http:// www. nat.rev.neurol.com